CLAIMS

1. Use of a composition comprising an isolated human truncated cystathionine beta synthase (htCBS) mutant polypeptide in the manufacture of a medicament for the treatment of homocystinuria in a subject, wherein the composition is to be administered to the subject, and wherein:

(a) the isolated htCBS mutant polypeptide comprises amino acids 2-413 of the amino acid sequence of SEQ ID NO:13 and has been PEGylated with an NHS ester-activated PEG molecule having a molecular weight greater than or equal to 20 kDa; and

(b) the isolated htCBS mutant polypeptide is to be administered in a dose between 0.33 mg/kg and 10 mg/kg at least once a week for at least six weeks.

2. The use of claim 1, wherein cystathionine and/or cysteine is increased in a subject.

3. The use of claim 1, wherein homocysteine, methionine, and/or S-adenosyl homocysteine is decreased in a subject.

4. The use of any one of claims 1-3, wherein the dose is an amount selected from the group consisting of: 0.33 mg/kg, 0.4 mg/kg, 0.5 mg/kg, 0.66 mg/kg, 1 mg/kg, 2 mg/kg, and 5 mg/kg.

5. The use of any one of claims 1-3, wherein isolated htCBS mutant polypeptide is to be co-administered with betaine.

6. The use of any one of claims 1-3, wherein betaine is to be administered prior to the isolated htCBS mutant polypeptide.

7. The use of claim 1 or claim 2, wherein the htCBS mutant polypeptide is to be administered at least twice a week.

8. The use of claim 2, wherein the amount of cystathionine is increased to above 0.008 µM or to between 0.005-0.35 µM.

9. The use of claim 8, wherein the amount of cysteine is increased to above 140 µM or to between 200 µM to 400 µM.

10. The use of claim 3, wherein the amount of homocysteine is decreased to less than 100 µM or to 10 µM.

11. The use of claim 10, wherein the amount of methionine is decreased to less than 50 μM or to 30 μM.

12. The use of claim 11, wherein the amount of S-adenosyl homocysteine is decreased to less than 0.14 μM or to 0.015 μM.